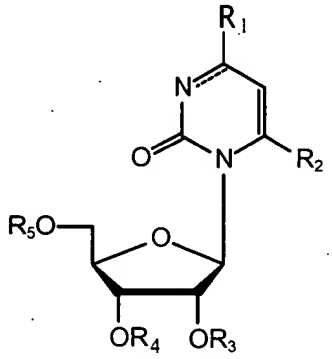
Exhibit B

CLAIMS UPON ENTRY OF THE AMENDMENT

1. A method for the treatment of a mitochondrial disorder comprising administering to a subject having or at risk of having such disorder an effective amount of a compound of Formula I:

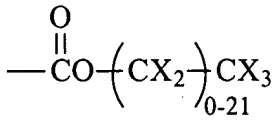


(I)

wherein:

R_1 is O, OH, $NHCOCH_3$, or NH_2,

R_2 is H, CO_2H, or



wherein:

each X is independently H or optionally substituted C_1-C_{22} alkyl, optionally substituted C_1-C_{22} alkenyl, or optionally substituted C_1-C_{22} alkynyl, with substituents selected from the group consisting of H, C_1-C_3 alkyl, OH, NH_2, and halogen,

R_3, R_4, and R_5 are each independently optionally substituted C_1-C_{22} alkyl carbonyl, with substituents selected from the group consisting of C_1-C_3 alkyl, OH, NH_2, and halogen, or H, wherein at least one of R_3, R_4, and R_5, are not H,

thereby treating the disorder.

2. A method for the treatment of a mitochondrial disorder comprising administering to a subject having or at risk of having such disorder, an effective amount of 2',3',5'-tri-*O*-acetyl-1-β-D-uridine.

3. A method according to claim 1 or claim 2, wherein the optionally substituted alkyl carbonyl is unbranched and has in the range of about 5 to 22 carbons.

4. A method according to claim 1, wherein the alkyl carbonyl is a carbonyl derivative of an amino acid selected from the group consisting of glycine, L-forms of alanine, valine, leucine, isoleucine, tyrosine, proline, hydroxyproline, serine, threonine, cystine, cysteine, aspartic acid, glutamic acid, arginine, lysine, histidine, carnitine, and ornithine.

5. A method according to claim 1, wherein the alkyl carbonyl is a carbonyl derivative of a dicarboxylic acid having in the range of about 3 to 22 carbons.

6. A method according to claim 1, wherein the mitochondrial disorder is a primary disorder comprising at least one mutation in mitochondrial or nuclear DNA.

7. A method according to claim 1 or claim 2, wherein the mitochondrial disorder is selected from the group consisting of Huntington's disease, Amyotrophic lateral sclerosis, MELAS (Mitochondrial encephalomyopathy with lactic acidemia and stroke-like episodes), MERRF (Myoclonus, epilepsy, and myopathy with ragged red fibers), NARP/MILS (Neurogenic muscular weakness, ataxia, retinitis pigmentosa/Maternally inherited Leigh syndrome), LHON (Lebers hereditary optic neuropathy) "Mitochondrial blindness", KSS (Kearns-Sayre Syndrome),

PMPS (Pearson Marrow-Pancreas Syndrome), CPEO (Chronic progressive external opthalmoplegia), Leigh syndrome, Alpers syndrome, Multiple mtDNA deletion syndrome, MtDNA depletion syndrome, Complex I deficiency, Complex II (SDH) deficiency, Complex III deficiency, Cytochrome c oxidase (COX, Complex IV) deficiency, Complex V deficiency, Adenine Nucleotide Translocator (ANT) deficiency, Pyruvate dehydrogenase (PDH) deficiency, Pyruvate carboxylase deficiency, Ethylmalonic aciduria with lactic acidemia, 3-Methyl glutaconic aciduria with lactic acidemia, Refractory epilepsy with declines during infection, Asperger syndrome with declines during infection, Autism with declines during infection, Attention deficit hyperactivity disorder (ADHD), Cerebral palsy with declines during infection, Dyslexia with declines during infection, MNGIE (Mitochondrial myopathy, peripheral and autonomic neuropathy, gastrointestinal dysfunction, and epilepsy), MARIAHS syndrome (Mitochondrial ataxia, recurrent infections, aphasia, hypouricemia/hypomyelination, seizures, and dicarboxylic aciduria), ND6 dystonia, Cyclic vomiting syndrome with declines during infection, 3-Hydroxy isobutryic aciduria with lactic acidemia, Diabetes mellitus with lactic acidemia, Familial Bilateral Striatal Necrosis (FBSN), Aminoglycoside-associated deafness, Dilated or hypertrophic cardiomyopathy, Wolfram syndrome, Multiple mitochondrial DNA deletion syndromes, and Renal Tubular Acidosis/Diabetes/Ataxia syndrome.

8. A method according to claim 1 or claim 2, wherein the mitochondrial disorder is a deficiency of cardiolipin.

9. A method according to claim 1 or claim 2, wherein the mitochondrial disorder comprises a deficiency in a pyrimidine synthetic pathway.

10. A method according to claim 9, wherein the deficiency in a pyrimidine synthetic pathway is the uridine synthetic pathway.

11. A method according to claim 9, wherein the deficiency comprises reduced expression and/or activity of an enzyme in the pyrimidine synthetic pathway.

12. A method according to claim 11, wherein the enzyme is selected from the group consisting of dihydroorotate dehydrogenase (DHOD) and uridine monophosphate synthetase (UMPS).

13. A method according to claim 1 or claim 2, wherein the mitochondrial disorder results in lower than normal uridine levels.

14. A method according to claim 1 or claim 2, wherein the mitochondrial disorder is the result of prior or concurrent administration of a pharmaceutical agent.

15. A method according to claim 14, wherein the pharmaceutical agent is a reverse transcriptase inhibitor, a protease inhibitor or an inhibitor of DHOD.

16. A method according to claim 15, wherein the reverse transcriptase inhibitor is Azidothymidine (AZT), Stavudine (D4T), Zalcitabine (ddC), Didanosine (DDI) or Fluoroiodoarauracil (FIAU).

17. A method according to claim 15, wherein the protease inhibitor is Ritonavir, Indinavir, Saquinavir or Nelfinavir.

18. A method according to claim 14, wherein the DHOD inhibitor is Leflunomide or Brequinar.

19. A method according to claim 1 or claim 2, further comprising the administration of one or more co-factors, vitamins, or mixtures of two or more thereof.

20. A method according to claim 19, wherein the co-factor is one or both of Coenzyme Q10 or calcium pyruvate.

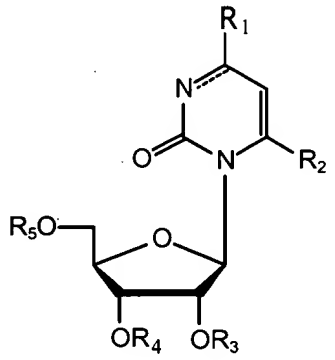
21. A method according to claim 19, wherein the vitamin is selected from the group consisting of thiamine (B1), riboflavin (B2), niacin (B3), pyridoxine (B6), folate, cyanocobalamine (B12), biotin, α-lipoic acid, and pantothenic acid.

22. A method according to claim 1, wherein the compound of Formula (I) is administered in a daily dosage in the range of about 0.5 g/m^2 to 20 g/m^2.

23. A method according to claim 1, wherein the compound of Formula(I) is administered in a daily dosage in the range of about 2 g/m^2 to 10 g/m^2.

24. A method according to claim 1, wherein the compound of Formula(I) is administered in a daily dosage of about 6.0 g/m^2.

25. A method for reducing or eliminating one or more symptoms associated with a mitochondrial disorder comprising administering to a subject in need thereof an effective amount of a compound of Formula I:

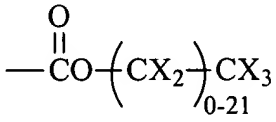


(I)

wherein:

R_1 is O, OH, $NHCOCH_3$, or NH_2,

R_2 is H, CO_2H, or



wherein:

each X is independently H or optionally substituted C_1-C_{22} alkyl, optionally substituted C_1-C_{22} alkenyl, or optionally

substituted C_1-C_{22} alkynyl, with substituents selected from the group consisting of H, C_1-C_3 alkyl, OH, NH_2, and halogen,

R_3, R_4, and R_5 are each independently optionally substituted C_1-C_{22} alkyl carbonyl, with substituents selected from the group consisting of C_1-C_3 alkyl, OH, NH_2, and halogen, or H, wherein at least one of R_3, R_4, and R_5, are not H,

thereby treating the disorder.

26. A method for reducing or eliminating one or more symptoms associated with a mitochondrial disorder comprising administering to a subject having or at risk of having such disorder, an effective amount of triacetyluridine.

27. A method according to claim 25 and claim 26, wherein said symptoms are renal tubular acidosis (RTA), impaired eyesight, dementia, seizures, cardiomyopathy, skeletal myopathy, peripheral myopathy or autonomic myopathy.

28. A method according to claim 1 or 2, wherein the mitochondrial disorder is selected from the group consisting of MELAS (mitochondrial encephalomyopathy with lactic acidemia and stroke-like episodes), MERRF (myoclonus, epilepsy, and myopathy with ragged red fibers), NARP/MILS (neurogenic muscular weakness, ataxia, retinitis pigmentosa/maternally inherited Leigh syndrome), LHON (Lebers hereditary optic neuropathy, "mitochondrial blindness"), KSS (Kearns-Sayre Syndrome), PMPS (Pearson Marrow-Pancreas Syndrome), CPEO (chronic progressive external opthalmoplegia), Leigh syndrome, Alpers syndrome, multiple mtDNA deletion syndromes, mtDNA depletion syndromes, complex I deficiency, ND6 dystonia, complex II (SDH) deficiency, complex III deficiency, cytochrome C oxidase (COX, complex IV) deficiency, complex V deficiency, adenine nucleotide translocator (ANT) deficiency, pyruvate carboxylase deficiency, and pyruvate dehydrogenase (PDH) deficiency.

29. A method for treating or preventing pathophysiological consequences of mitochondrial respiratory chain dysfunction in a mammal comprising administering to said mammal in need of such treatment or prevention an effective amount of a pyrimidine nucleoside.

30. A method as in claim 29 wherein said respiratory chain dysfunction is caused by a mutation, deletion, or rearrangement of mitochondrial DNA.

31. A method as in claim 29 wherein said respiratory chain dysfunction is a deficit in mitochondrial Complex I activity.

32. A method as in claim 29 wherein said respiratory chain dysfunction is a deficit in mitochondrial Complex II activity.

33. A method as in claim 29 wherein said respiratory chain dysfunction is a deficit in mitochondrial Complex III activity.

34. A method as in claim 29 wherein said respiratory chain dysfunction is a deficit in mitochondrial Complex IV activity.

35. A method as in claim 29 wherein said respiratory chain dysfunction is a deficit in mitochondrial Complex V activity.

36. A method as in claim 29 wherein said pyrimidine nucleotide is administered orally.

37. A method as in claim 29 wherein said pathophysiological consequence of mitochondrial respiratory chain dysfunction is a congenital mitochondrial disease.

38. A method as in claim 37 wherein said congenital mitochondrial disease is selected from the group consisting of MELAS, LHON, MERRF, MNGIE, NARP, PEO, Leigh's Disease, Alpers syndrome, mitochondrial cytopathy, mitochondrial myopathy, mitochondrial encephalomyopathies, and Kearns-Sayre Syndrome.

39. A method as in claim 29 wherein said pathophysiological consequence of mitochondrial respiratory chain dysfunction is a neurodegenerative disease.

40. A method as in claim 39 wherein said neurodegenerative disorder is Alzheimer's Disease.

41. A method as in claim 39 wherein said neurodegenerative disorder is Parkinson's disease.

42. A method as in claim 39 wherein said neurodegenerative disorder is Huntington's Disease.

43. A method as in claim 29 wherein said pathophysiological consequence of mitochondrial respiratory chain dysfunction is selected from the group consisting of renal tubular acidosis, dilating or hypertrophic cardiomyopathy, steatohepatitis, hepatic failure, and lactic acidemia.

44. A method for treating developmental delay in cognitive, motor, language, executive function, or social skills in a mammal comprising administration of an effective amount of a pyrimidine nucleoside.

45. A method as in claim 44 wherein said developmental delay is a subset of Attention Deficit/Hyperactivity Disorder.

46. A method as in claim 44 wherein said developmental delay is a subset of autism associated with mitochondrial dysfunction.

54. A method as in claim 29 wherein said pyrimidine nucleoside is selected from the group consisting of uridine, cytidine, an acyl derivative of uridine, an acyl derivative of cytidine, orotic acid, an alcohol ester of orotic acid, or a pharmaceutically acceptable salt thereof.

55. A method as in claim 54 wherein said pyrimidine nucleoside is administered orally.

56. A method as in claim 29 wherein said pathophysiological consequence of mitochondrial respiratory chain dysfunction is a congenital mitochondrial disease.

57. A method as in claim 56 wherein said congenital mitochondrial disease is selected from the group consisting of MELAS, LHON, MERRF, NARP, PEO, Leigh's Disease, Alpers syndrome, and Kearns-Sayre Syndrome.

62. A method as in claim 29 wherein said pathophysiological consequence of mitochondrial respiratory chain dysfunction is selected from the group consisting of renal tubular acidosis, dilating or hypertrophic cardiomyopathy, steatohepatitis, hepatic failure, and lactic acidemia.

66. A method according to claim 1, wherein said mitochondrial disorder is a secondary disorder caused by acquired somatic mutations, physiologic effects of drugs, viruses, or environmental toxins that inhibit mitochondrial function.